

## LATE CONGENITAL SYPHILITIC NERVE DEAFNESS\*†

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Ritchie Rodger (1945) speaking to this Society classified congenital syphilitic nerve deafness into early and late. He gave two varieties of the early type; a neuritis associated with basal meningitis, and an oto-labyrinthitis. These two early varieties were found to occur in infancy. There is only one late type, a labyrinthitis, which is by far the commonest of all varieties; this usually occurs after the age of 9 years, but may appear at any period in adult life. The following remarks apply to this late type only.

The exact nature of the pathology of late syphilitic nerve deafness has never been settled. There is still some support for the idea that the condition is caused by degeneration in the eighth nerve itself. Degeneration of midbrain nuclei or pressure from inflammatory changes around the nerve have also been suggested. *Post-mortem* material is, by the nature of the condition, not readily available. Apart from a few individual reports there is only one collected series in the literature. Mayer and Fraser (1936) described a serous labyrinthitis which they believed to be consequent on periostitis and osteitis in the bone surrounding the cavities and canals of the internal ear. Microscopically, they believed the lesions to be miliary gummata. There is no report that a search was made for *Spirochaeta pallida*. This work has never been confirmed. It is interesting to note that, although the patients described had had treatment, new and healed microscopic lesions existed side by side.

Clinically, late congenital syphilitic nerve deafness is often total and of sudden origin. In others it starts mildly and progresses rapidly or slowly. Usually it is bilateral and is much commoner in females. It often follows the occurrence of interstitial keratitis by some 2 or 3 years, or even longer. In association with the deafness many patients complain of tinnitus, which may precede or follow

deafness and be persistent. Associated vertigo is not uncommon. These patients usually have a strongly positive blood Wassermann, and invariably a normal cerebrospinal fluid. The condition appears to be uninfluenced by antisyphilitic treatment, and indeed deafness may occur during therapy, and even after what is generally regarded as adequate treatment.

Some of the less dramatic cases bear a resemblance to Menière's disease, in which vasodilator drugs have recently been used with varying success (Fisher and Tebrock, 1953). The reports give the impression that the exhibition of vasodilator drugs is worth a trial in every case. Menière's disease is another condition in which the pathology is obscure. The consensus of opinion favours a labyrinthitis of vasomotor origin (Brunner, 1948), the vasomotor changes being due to allergy or hypersensitivity to some toxin which may be metabolic or infective in origin. It will be remembered that Menière's disease is manifest by progressive deafness, associated with attacks of tinnitus and vertigo. The deafness usually worsens after each of the attacks, and in not a few cases the patient progresses to total bilateral nerve deafness.

Is it possible that these two conditions, late syphilitic congenital nerve deafness and Menière's disease, are in some way related? Is it possible that they have a common type of origin in hypersensitivity of some kind? If this were so, late congenital syphilitic nerve deafness would be placed in the same category as interstitial keratitis. This would explain in some measure the failure of the condition to respond to antisyphilitic treatment, and its appearance or progress in spite of such treatment. On these very theoretical grounds it was considered that a trial with vasodilator drugs might be worth while in syphilitic nerve deafness.

Ronicol,\* the alcohol of nicotinic acid, the active principle being beta-pyridyl carbinol, was the drug used. Ronicol produces gradual and prolonged vaso-

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\* "Ronicol" is manufactured by Roche Ltd.

dilatation, has very few side-effects, and may be administered for long periods. It is made up in 25-mg. tablets. Four cases of late congenital syphilitic nerve deafness are described below.

#### Case Reports

**Case 1, a female, aged 53,** first attended in February, 1952, with iridocyclitis. The Wassermann reaction was strongly positive and the cerebrospinal fluid normal. There was no history or evidence of acquired syphilis. The husband and three children were Wassermann reaction negative, and the patient stated that her mother had been fully treated for syphilis. The patient gave a history of bilateral nerve deafness with head noises for only a few months before her initial attendance. The condition had started with head noises, followed 3 weeks later by the onset of deafness. She was examined by an aural specialist, who diagnosed congenital syphilitic nerve deafness. Full antispecific treatment was completed some 2 years after her initial attendance. There was no improvement in the deafness. Trial of Ronicol was started in November, 1954, using 25 mg. three times a day. At the end of a fortnight there was no change. The dosage was increased to 50 mg. three times a day, and after a further 2 weeks of this treatment the patient stated that the deafness was about the same, although occasionally voices sounded "very tinny". The head noises were no better, but she stated that her attacks of dizziness were slightly less frequent. Ronicol treatment was discontinued.

**Case 2, a female, aged 22,** first attended in August, 1952, with antenatal Wassermann reaction positive. There was a history of interstitial keratitis in 1943, when she received full and prolonged antispecific treatment. Deafness commenced in 1948 and progressed rapidly. From the beginning it was associated with severe head noises, and difficulty in balancing, especially in the dark. There were no neurological signs, and the cerebrospinal fluid was normal. Apart from an "insurance" course of penicillin during the pregnancy, she had no further antispecific treatment. Ronicol 25 mg. three times a day was started in October, 1954, and continued for 14 days. For the first few days the head noises were very much worse, and the patient had great difficulty in getting to sleep. Thereafter the condition settled down, and at the end of a month, in spite of increasing the dosage, no change was noted.

**Case 3, a female, aged 40,** whose mother had died of general paralysis of the insane. There were no other members of the family. She first attended in 1947 with a gummatous ulcer of the left shoulder and received about two courses of antispecific treatment before defaulting. Bilateral deafness, associated with severe head noises and vertigo, first started in 1949 and advanced rapidly for 3 weeks. In this case a hearing aid was of little value in view of the severe head noises. Ronicol 25 mg. three times a day was started in October, 1954. After a few days, the patient complained of flushing of the face. There was no change in the deafness,

vertigo was much the same, but she stated that the head noises were "not so bad".

In view of the face flushing, the dosage was reduced to  $\frac{1}{2}$ -tablet three times a day, but the side-effects continued to be troublesome, and, although the tinnitus improved further, treatment was discontinued. With the discontinuance of treatment, the head noises became as bad as previously. Later, she asked for further supplies of the tablets, as she was in no doubt that they helped the head noises and made her hearing aid more useful.

**Case 4, a female, aged 35,** first attended in April, 1952, with antenatal Wassermann reaction positive. She had Hutchinsonian incisors. She received two courses of penicillin and one of bismuth, and was transferred to care elsewhere after the birth of the baby; 16 months after her original attendance she was referred back by an aural specialist with a diagnosis of congenital syphilitic nerve deafness, which had started 4 months previously. The patient had severe head noises, which seemed to be getting worse. There were also attacks of dizziness. Full antispecific treatment was completed in September, 1954, without any change in the patient's symptoms. In October, 1954, Ronicol treatment was started (25 mg. three times a day for 14 days). There was immediate improvement in the head noises, no change in the dizziness, but the patient thought her hearing had improved slightly, probably because the head noises had subsided. There were no side-effects, and the dosage was increased to 50 mg. three times a day. After a further fortnight the head noises and the dizziness were giving her no trouble. There was definite and sustained improvement in the hearing. She asked for further supplies of Ronicol, and has continued taking the drug for 6 months. Tinnitus and vertigo have remained completely absent, and she has discontinued using her hearing aid and states that her hearing has improved.

#### Discussion

This trial series is very small, and no firm conclusion can be based on the findings. Apart from the fact that all the patients admitted to some change in symptomatology while taking Ronicol, only one showed changes which could be classified as improvement. It is interesting to note that this case is the one of most recent origin. Further trials would seem worth while especially in early cases.

It may well be that the more recent the history of deafness the greater the hope of improvement.

This raises the point that, so far, we are not able to tell which cases of congenital syphilis are liable to develop nerve deafness. The pathological changes described by Mayer and Fraser (1936) suggest that bony changes may be present for some years before the onset of symptoms. If we can find a method of detecting pathological changes in the ear before deafness has occurred we may not only be in a position to differentiate between congenital and latent syphilis but also may have an opportunity

to try to prevent deafness. If the nerve deafness is a result of syphilitic periostitis with labyrinthitis, the microscopical changes being brought about by the presence of spirochaetes in the internal ear, then one may consider the use of vasodilator drugs as a useful adjunct to antisyphilitic treatment, that is, with a view to concentrating drugs, for example, penicillin, at the site of the inflammation. If, however, late congenital nerve deafness and interstitial keratitis are manifestations of the same hypersensitivity, and akin to Menière's disease, then one will have to consider whether vasodilator drugs are likely to be useful, or whether cortisone, or cortisone-like treatment, *e.g.*, fever therapy, or prolonged aspirin treatment, should be tried.

Congenital syphilis is becoming less common. We have, however, to deal with a back-log of cases. It is likely that some 10 to 15 per cent. of the late congenital syphilitics will present with, or subse-

quently develop, nerve deafness. It is now nearly 100 years since Hutchinson (1863) described his triad. The notched incisors which he described have never been a problem. If found aesthetically unacceptable to the patient (or her medical attendant), they can be extracted and replaced by a prosthesis. Interstitial keratitis we can now control almost completely with topical cortisone. The time seems ripe to turn our attention to the third component of the triad, and these few thoughts on the subject are, therefore, advanced for consideration.

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